International Journal of Healthcare Sciences ISSN 2348-5728 (Online)
Vol. 2, Issue 2, pp: (164-166), Month: October 2014 - March 2015, Available at: www.researchpublish.com

A Case Of Extra-Cranial Shoulder Arterio-Venous Malformation And Its Management Challenge; Case Report

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Abstract: **Arteriovenous malformations (AVM) occurring extra-cranially are rare compared to their intracranial counterpart. The potential for expansion of these AVMs throughout life with soft tissue invasion, bleeding, deformity, and functional deficits has led to much concern in terms of management. We report a case of shoulder AVM in a boy and the management dilemma it poses when he presented with multiple bleeding episodes from the ulcerated mass. A 13-year-old boy was referred for bleeding due to an infected right shoulder ulcer for 3 years. Angiogram done previously showed a right shoulder AVM. There was no bleeding, pain or ulceration at that time. Embolization was done twice. However, he was admitted with recurrent episodes of bleeding managed by repeated embolization. We were unable to completely identify/embolise all feeding vessels to the AVM. His wound remained dirty, easily bleed during dressing change and he required repeated blood transfusions. A multidisciplinary team was involved, including vascular surgeons, interventional radiologists, plastic surgeons, paediatric orthopaedic surgeons and an orthopaedic oncologic surgeon. Management options were discussed. It was clear that excision was not feasible as it extends to the neck region. A conservative option in the form of "embolise if bleed" seemed to be the most feasible option.**

Keywords: **Arteriovenous malformations; angioembolization; extra-cranial; shoulder.**

I. INTRODUCTION

Arteriovenous malformations (AVM) are high-flow lesions having a direct connection between an artery and vein, with bypass of the capillary bed. Those affecting the upper limb and causing cardiac decompensation are rare. [1] A thorough clinical examination and history can usually establish the diagnosis of hemangioma or vascular malformation. [2] Before invasive treatment for vascular malformations, detailed imaging diagnostic procedures such as catheter angiography are required. [3] Color Doppler imaging (CDI) is an essential tool in the diagnostic evaluation of vascular malformations. Both high-flow lesions (AVMs, AVFs) and low-flow lesions (venous malformations, lymphatic malformations) can be diagnosed accurately. [2]

Computed tomography (CT), although helpful in the diagnostic evaluation, is less useful than magnetic resonance imaging (MRI). Unlike CT, MRI easily distinguishes between high-flow and low-flow vascular malformations. Furthermore, the anatomic relationships of the vascular malformation to adjacent nerves, muscles, tendons, solid organs, bone, and subcutaneous fat allow a complete assessment. MRI is also an excellent noninvasive method for observing patients to determine the efficacy of therapy, often obviating repetitive arteriography and venography. CT has its main role in intraosseous vascular malformations and determination of the extent of bone involvement. [2]

Vascular malformations were initially treated by surgeons alone. The early rationale of proximal arterial ligation of arteriovascular malformations (AVMs) proved totally futile as the phenomenon of neovascular recruitment reconstituted arterial inflow to the AVM nidus. Microfistulous connections became macrofistulous feeders. Complete surgical extirpation of a vascular malformation can be very difficult and, at times even hazardous, necessitating suboptimal partial resections. Partial resections can cause a good initial clinical response that may last for some time. [2] It was reported that it only 20% of malformations may be amenable to complete extirpation with surgery. [2] For some patients, limb amputation becomes necessary and carries significant risk. [1] Not only the prognosis, but also the treatment methods for vascular malformations vary according to the particular lesions of each patient. [3]

II. CASE REPORT

13 year old boy, was referred from Kuantan Hospital for further management for bleeding from infected right shoulder ulcer with underlying arteriovenous malformation (AVM) over the right shoulder since 2009 (Fig. 1). He gave history of recurrent fever and murmur which revealed compensated heart failure. Angiogram was done in 2004, diagnosed to have AVM of right shoulder; there was no bleeding, pain or ulceration at the time. Embolization done twice, in 2004 and 2007 in another tertiary centre. However, recurrent episodes of bleeding occurred, but managed by re-embolisations, but unable to completely identify or embolise all feeding vessels. Wound remains dirty, easily bleeds during dressing change, and requires repeated blood transfusions. MRA not done due to metal coils causing artifact. So, a multidisciplinary team discussion composed of vascular Surgeons, interventional radiologists, plastic surgeon, paediatric orthopaedic surgeon and an orthopaedic oncologic surgeon. Treatment options were discussed in details. As there were multiple feeding vessels involving vertebral artery, intercostal artery, subclavian artery and external carotid artery (Fig. 2. A, B and C). It was obvious that excision was not feasible as the lesion extends to neck region, also to excise and raise a skin flap there is a likelihood of flap necrosis. Subclavian brachial bypass or ligation of feeders, is a major surgery with unsure of outcome. Radiation will worsen ulceration, so, excluded. Endovascular stenting via angiogram with or without embolisation of remainder feeding vessels was another option. Conservative watchful managment with "embolise if bleeds" plan seemed the only reasonable plan.

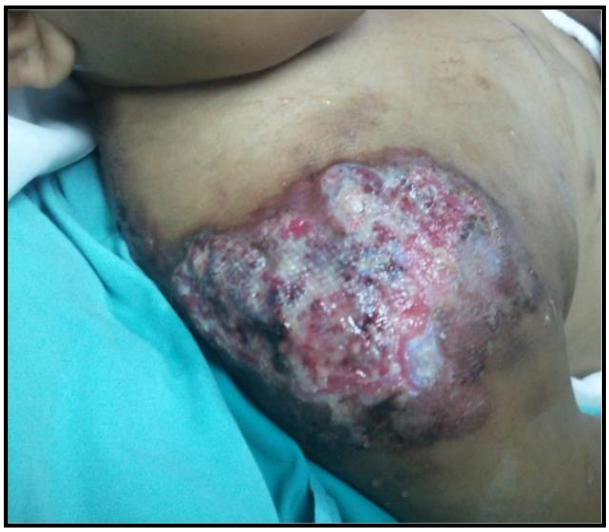

Fig. 1. AVM with overlying skin, which shows chronic ulceration with stigmata of recent bleed and infection

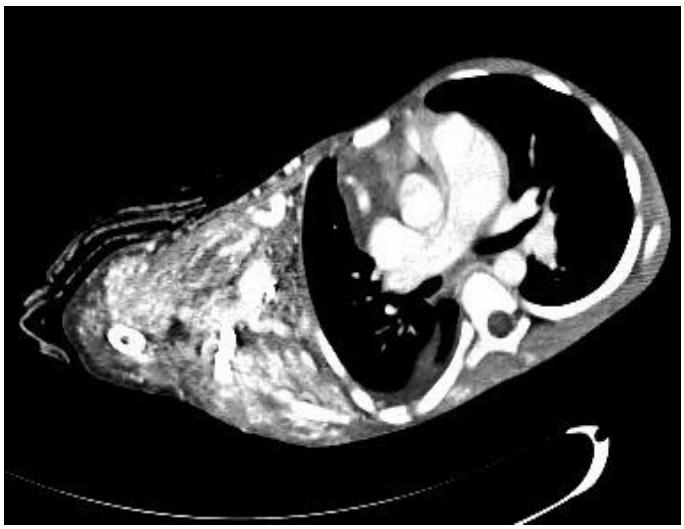

Fig. 2. A. CT-Angiogram of the shoulder shows complex AVM with multiple feeders from chest wall and root of the neck.

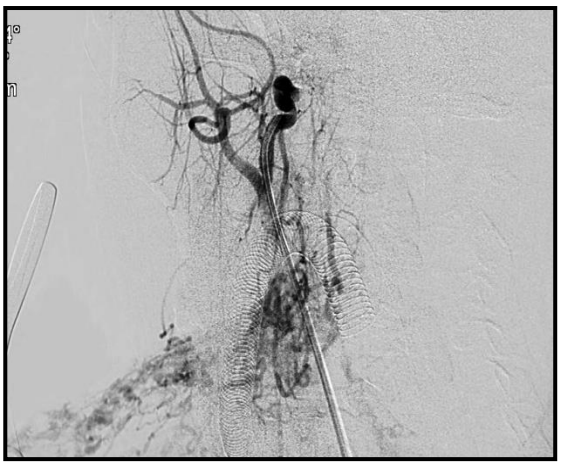

Fig. 2. B. Angiogram of the AVM during embolization of one of the feeders with evidence of previous coiling

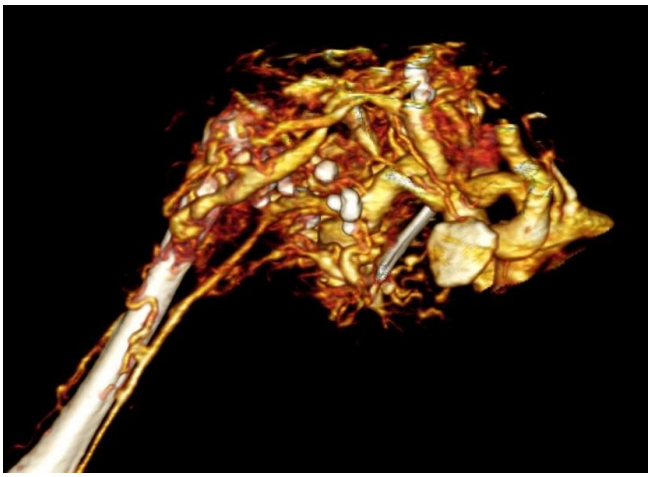

Fig. 2. C. Reconstructed CT-Angiogram of the shoulder shows the complexity of the AVM

III. DISCUSSION

AVM is a congenital abnormal connection between arterial and venous systems mediated by abnormal capillaries (a nidus). AVMs present at birth deteriorate slowly with age. [3] Clinical presentation of upper limb AVMs can be diverse. Patients may suffer pain, nerve compression, ulceration, hand dysfunction, and spontaneous bleeding. Shunting through proximal arteriovenous fistulae may result in distal steal phenomenon manifesting as severe pain, ischaemia, and discoloration of the digits. [4] The presence of an upper-limb AVM does not correlate with vascular anomalies of other organ systems, nor does the pattern predict skeletal overgrowth. [5]

The management of AVM remains a significant challenge, particularly those of the upper limb. Intervention should only be contemplated when the lesion becomes symptomatic. A variety of treatment strategies have been reported with varying success. [1] Management of AVM varies, with a conservative approach adopted for patients that are asymptomatic or have minor symptoms. If treatment is required, techniques that may be used include catheter embolization (our case) or direct percutaneous sclerotherapy. Embolization and sclerotherapy may be performed even after major surgery. [6]

Regardless of the embolic agent used, the underlying aim is to obliterate the nidus. However, embolotherapy must not be considered a definitive cure and can unintentionally worsen symptoms. Furthermore, repeat embolization may be necessary to deal with recurrences. [1] More invasive treatments might be required for patients with vascular malformation if they have uncontrollable pain, clinically significant heart strain, intractable ulcer, functional disorder, or disfigurement. Either complete resection or occlusion of the nidus is required for complete cure of AVM. [3] Therefore, in many cases, the aim of treatment for vascular malformation is for symptomatic relief rather than a cure. [7]

A multidisciplinary approach is central to the effective management of AVMs. These lesions are complex and difficult to treat. Despite advances in embolic agents and microcatheter techniques, outcomes for high-flow upper-limb AVM is far from satisfactory. For diffuse lesions that are life-threatening, amputation as the first step in the patient's rehabilitation may be appropriate. [1]

IV. CONCLUSION

AVMs pose a major challenge to treat in medical practice. It is often associated with serious symptoms such as heart failure, neuropathy, pain, and bleeding. Small and superficial AVMs can be cured with surgical resection. However most AVMs are inoperable, being large and diffuse, involving important normal adjacent structures. If left untreated patients are at risk of hyperdynamic cardiac failure and uncontrolled bleeding episodes. Vascular malformations are best treated where a team is in place that regularly sees patients with these malformations. Only then can rational decisions and advancements in treatments occur.

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